Virilizing adrenal carcinoma with inferior vena cava thrombus

Sir

Virilizing adrenal carcinoma is a very rare disease. It occurs with an estimated incidence of one per 1.7 million. Although rare, this tumor resembles renal cell carcinoma in its propensity to develop venous thrombus. Aggressive surgical therapy is required as there is no reliable adjuvant therapy. This case of adrenal cortical

carcinoma with virilizing symptoms along with inferior vena cava (IVC) thrombus is not reported in literature. A 54-yearr-old female presented with a complaint of pain in her left upper abdomen. She complained of hirsutism for last three months along with baldness. On examination her pulse was 84/minute and blood pressure was 160/90 mmHg. Facial hair growth was present with temporal balding. Abdominal examination revealed increased hair growth in the midline area. A lump of size 12 x 10 cm was present in the left hypochondrium, extending to the left lumbar region.

Renal function, liver function tests, and serum electrolyte were normal. Urinary vanillylmandelic acid was 3.2 mg / 24 hours (normal up to 7 mg / 24 hours). Serum testosterone was 248 ng/dl and Dehydroepiandrosterone (DHEA) sulfate was 420 g / dl. Serum cortisol was 14 mg / dl. A contrast CT of the abdomen revealed a heterogeneous solid mass of 11.7 x 9.5 cm in the suprarenal region, separate from the upper pole of the left kidney and splenic parenchyma, and an IVC thrombus was present up to the hepatic veins [Figure 1]. Surgery was performed with enblock resection of the left suprarenal mass along with the removal of the kidney and IVC thrombus on venovenous bypass.

On histology the tumor had a high nuclear grade (Fuhrman 3), necrosis, atypical mitosis, and venous invasion. Postoperative recovery was uneventful. The patient was followed up with hormonal assays and had a satisfactory recovery

Adrenal cortical carcinoma has bimodal age distribution, with its peak in the first and fifth decade. It is more common in women than in men, and its presentation as virilization is very rare. These functioning Androgen producing tumors cause virilization and marked elevations of serum testosterone and dehydroepiandrosterone.[1,2]

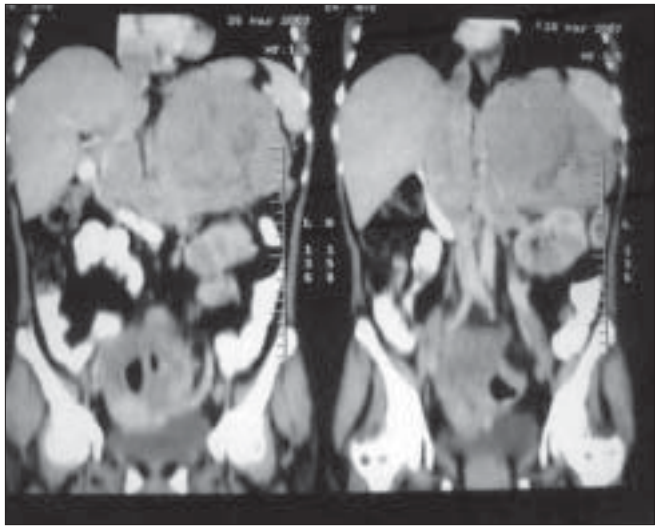

Figure 1: CT-scan showing heterogeneous solid mass in the left suprarenal region involving kidney and IVC

These tumors often extend directly into the adjacent structures, especially the kidney, and may involve the inferior vena cava. The first reported case of vena caval tumor thrombus involvement was described intraoperatively by Castleman *et al*.[3] Vena caval tumor thrombus involvement can occur either by direct extension or more commonly by intraluminal extension of the tumor thrombus via the adrenal or renal vein. Many authors have seen a propensity of right-sided tumor thrombus extensions, which may be attributed to a short right adrenal vein.[2] The patient in our case had a left-sided tumor that had extended intraluminally via the adrenal and renal veins.

With the advent of newer imaging techniques most of the adrenal masses are detected in silent form (1 to 10%). The estimated prevalence of silent adrenal cortical carcinoma is less than 1 in 250,000.[1] Adrenal malignancies are usually larger than 6 cm. Belldegrun and coworkers reviewed six series, and found that 105 of 114 adrenocortical carcinomas were larger than 6 cm[4] From a metabolic standpoint, 17-ketosteroid and DHEAS levels are often high in patients with carcinomas, usually in conjunction with elevated glucocorticoid production.[5] In our case the size of the tumor was more than 6 cm and had elevated DHEAS and serum testosterone, it was labeled as a malignant lesion. Weiss proposed nine histological criteria; if more than three were present the lesion was considered malignant. In our case the tumor had four features.[6]

An abdominal CT is the standard radiographic method for evaluating a primary tumor, adjacent organ involvement, and for evaluating IVC thrombus. Magnetic Resonance Imaging (MRI) currently offers the most accurate preoperative technique. Percutaneous fine needle aspiration cytology under computed tomography (CT) on USG-guided aspiration biopsy is unreliable in preoperative diagnosis, as it is unable to differentiate adrenal adenoma from carcinoma. Adrenal Cortical Carcinoma (ACC) is generally an aggressive tumor and prognosis of adrenal cortical carcinoma is poor if untreated, with 20% survival at one year. The presence of distance metastasis reduces the tumor's five-year survival rate from 50 to 0%.[1] Surgical resection is the treatment of choice.

Combination chemotherapy can be used in an inoperable, recurrent, and metastatic disease.[1] Adjuvant therapy with mitotane has been used in patients with localized, completely resected disease, but its efficacy is questionable.[5]

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DOI: 10.4103/0019-509X.52966

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